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CLAUDE SYNDROME: A CASE REPORT

General Medicine

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ABSTRACT

Claude's syndrome caused by ventromedial midbrain lesion is characterized by ipsilateral third nerve palsy and contralateral ataxia. This syndrome is very rare and only a few cases have been reported since 1912. An 81 years old male developed sudden onset right third cranial nerve palsy and left sided ataxia. Magnetic resonance imaging (MRI) brain revealed an infarction in the right ventromedial midbrain. Although the red nucleus has often been suggested as the lesion site responsible for Claude's syndrome, a lesion of the superior cerebellar peduncle just below and medial to the red nucleus could be responsible for this syndrome.

KEYWORDS

cerebellar peduncle, Claude's, Magnetic resonance imaging, syndrome

INTRODUCTION

Midbrain lesions may give rise to complex eye movement disorders [1] .Three main types of dysfunctions are delineated. First, a fascicular syndrome of the third nerve is indicated when the peripheral type of third cranial nerve palsy is associated with some specific neurological disturbance (syndrome of Weber, Benedict, or Claude). Second, a nuclear syndrome is suggested when bilateral ptosis, bilateral mydriasis, bilateral or contralateral superior rectus weakness are present. And, third, a supranuclear syndrome usually includes vertical gaze palsy, skew deviation, sea-saw nystagmus, and vertical one-and-a half syndrome.

In 1912, a French neurologist Henri Claude first described the syndrome that bears his name[2]. His patient was a house painter who developed right third cranial nerve palsy with contralateral gait ataxia[3].The pathological examination revealed a paramedian mesencephalic infarction on the right involving the superior cerebellar peduncles, the medial half of the red nucleus, and some reports describe the syndrome with Claude's original red nucleus involvement[4-5]. The most common cause of Claude's syndrome is cerebrovascular disease and malignancy[6].

Detailed description of this syndrome is rare and there have been meagre studies on the imaging findings of the site of lesion involved in Claude syndrome. Only a few cases have been reported since 1912[7-9]. A patient presenting with clinical features and imaging findings suggestive of Claude's syndrome is described herein.

CASE REPORT

On March 15th, 2019, an 81 years old male was admitted for sudden onset of dizziness and inability to open his right eye. Before the onset of these symptoms, he was in good health except for uncontrolled hypertension and diabetes for past 25 years. He was taking an afternoon nap in a park when on waking up, he noted unsteadiness of gait while standing and had a tendency to fall towards left while attempting to walk. He had walked to the park all by himself while after the incident had to be brought back home with the help of attendants. Also, he developed sudden onset double vision that progressed over next four to six hours to complete drooping of upper eyelid of right eye.

There was no history of associated pain, mass or swelling over right eye. It was not accompanied by headache or facial deviation, numbness over face, nasal regurgitation, dysphagia or dysarthria. He did not complain of weakness over face or limbs or bladder/ bowel symptoms or abnormal involuntary body movements. There was no history of fever or malaise or preceding viral illness.

On examination, he was alert, conscious and oriented to time, place and person. BP was 160/100 mm of Hg. On neurological examination higher mental functions were normal. His gait was wide-based, and he swerved to the left when attempting to walk. Cranial nerves examination revealed pupil sparing right 3rd nerve palsy. Other cranial nerves were normal. Sensory system – Loss of vibration sense over limbs with position and joint sense impaired bilaterally, rest normal. Motor system- Nutrition and tone normal; power of grade 4+ in all limbs with no involuntary movements. Also, cerebellar signs were present on left that is finger nose, dysdiadochokinesia/ heel to knee test. There were no meningeal signs. Plantars were bilaterally flexor. Reflexes were normal. Fundus examination was suggestive of bilateral non- proliferative diabetic retinopathy.

INVESTIGATIONS

Fasting blood sugar level was 250 mg/dl. The Liver, renal function, lipid study, complete blood count, urinalysis, coagulation profile, and electrolyte test results were all within reference ranges. Electrocardiographic and echocardiographic findings revealed no cardiac source of emboli. The HbA1C and Urine albumin creatinine ratio were found to be 11% and 44 respectively. NCV was suggestive of distal symmetrical sensorimotor neuropathy in all four limbs. T2 weighted Magnetic Resonance Imaging(MRI) Brain, revealed an acute infarct in midbrain in right parasagittal area. MRI Orbit was normal. MR Angiogram was normal.

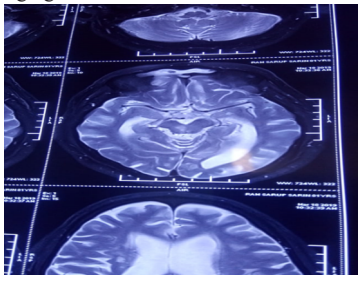

Figure 1: T2 weighted MRI Brain showing a high signal intensity lesion in the right ventromedial midbrain.

Figure 2: Right sided complete ptosis in our patient

DISCUSSION

Monocular palsy of elevation, depression, and adduction associated with complete ptosis in the right eye may correspond to involvement of the third cranial nerve fascicle or nucleus. Due to sparing of the pupil constrictor and contralateral lid elevator, nuclear involvement of the third cranial nerve was unlikely. The clinical feature of complete ipsilateral oculomotor nerve paralysis and contralateral cerebellar ataxia is consistent with Claude's syndrome that was further confirmed on imaging. He was started on aspirin, atorvastatin, enalapril and subcutaneous insulin (mixtard regimen). Also, supportive measures like gait training and physiotherapy were started.

The principal localization of Claude's syndrome has previously been attributed to a lesion of the red nucleus, since the cerebellar efferent fibres (dentatothalamic fibers) and oculomotor nerve fascicles meet at the level of the red nucleus [9]. Dentatothalamicfibers do not pass through the red nucleus, but ascend directly toward the lateral thalamus, sending a few branches to the red nucleus. Fibers of the superior cerebellar peduncle are found medial to the red nucleus at the level of the caudal end of the red nucleus. Only a few reported cases indicated additional lesions in the red nucleus [7]. In Claude's syndrome, the oculomotor nerve fascicles, as well as the cerebellar efferent fibers, are involved concurrently. At the red nucleus level, the oculomotor nerve fascicles do not meet the cerebellar efferent fibers; since the oculomotor nerve fascicles pass medially and the dentatothalamicfibers ascend laterally to the red nucleus. However, at the level of the caudal end of the red nucleus, the oculomotor nerve fascicles traverse the midbrain divergently and run across the superior cerebellar peduncle. Hence, a small lesion at this level can produce all the symptoms of Claude's syndrome.

Our patient had strictly unilateral oculomotor palsy and contralateral ataxia without involvement of the ipsilateral fourth nerve, opposite eye, or of sensory system. The pupil was also spared unlike the original description of the syndromes of Claude and Nothnagel. Our case is unique, in that it was a limited paramedian mesencephalic infarct involving superior cerebellar peduncle and third nerve fascicle.

CONCLUSION

Our patient presented with right pupil sparing oculomotor nerve palsy and left cerebellar ataxia suggestive of Claude's syndrome. The MRI findings revealed an acute infarct in right paramedian mesencephalon further confirming the diagnosis.

DECLARATION OF PATIENT CONSENT

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patient understands that his names and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Nil.

CONFLICTS OF INTEREST

There are no conflicts of interest.

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